

Case Report

Severe Daily Headache as an Uncommon Manifestation of Widespread Skull Base Osteomyelitis

Jens van der Valk^a Frank Treurniet^b Jan Pieter Koopman^c
Hille Koppen^d

^aLeiden University Medical Centre, Leiden, The Netherlands; ^bDepartment of Radiology, Haga Hospital, The Hague, The Netherlands; ^cDepartment of Ear, Nose and Throat, Haga Hospital, The Hague, The Netherlands; ^dDepartment of Neurology, Leiden University Medical Centre/Haga Hospital, Leiden, The Netherlands

Keywords

Headache · Secondary headache · Ear · Skull base · Infection

Abstract

Temporal bone osteomyelitis has been recognized for decades as a complication of otitis externa, specifically in elderly patients with diabetes. A much less prevalent form is skull base osteomyelitis. We report a 70-year-old man with diabetes who presented to our outpatient clinic with severe chronic daily complaints of headache. The headache was located frontoparietally and kept him awake at night. Imaging (nonenhanced computed tomography [CT], magnetic resonance imaging, and positron emission tomography/CT) showed a hypermetabolic mass on the right side of the skull base, in the middle ear, and in the mastoid process, with invasion and partial destruction of the surrounding elements of the petrous bone, the occipital bone, and the sphenoid bone on the right, with extension by way of the clivus into the apex of the left petrous bone. Diagnostic puncture revealed *Streptococcus pneumoniae*. The final diagnosis was severe daily headache due to central skull base osteomyelitis. Our case emphasizes the need for proper clinical and radiological investigation keeping the diagnosis of skull base osteomyelitis in mind with patients with diabetes or otherwise immunocompromised status who present with chronic daily headache and otalgia.

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Introduction

Temporal bone osteomyelitis has been recognized for decades as a complication of otitis externa, specifically in patients with diabetes, *Pseudomonas aeruginosa* being the usual pathogen [1]. Skull base osteomyelitis can be a complication of temporal bone osteomyelitis. Atypical skull base osteomyelitis is located in the sphenoid and/or occipital bones without associated external otitis [2]. We present a patient with severe chronic daily headache due to atypical skull base osteomyelitis.

Case Report

A 70-year-old man presented to our outpatient clinic because of headache complaints, which had been present for 6 months. The headache was located frontoparietally and was present continuously. The pain was pounding and kept him awake at night. Body posture did not influence the pain, whereas shaking could provoke the pain. The pain was rated 10 on a visual analogue scale. He also reported complaints of right otalgia for 9 months. No over-the-counter painkillers were effective. There were no signs of nausea or vomiting, although he reported photophobia. Autonomic symptoms were absent. He reported pain when combing his hair, but he had no jaw claudication. He had a history of diabetes, hypertension, deep vein thrombosis, and atrial fibrillation. He used oral anticoagulation. He reported no previous history of headaches.

The results of the neurological examination were normal, but the skin over the skull was sensitive. No papilledema was seen. Blood pressure and temperature were normal. Within 7 days, a computed tomography (CT) scan of the head was performed and showed a lytic lesion of the skull base; the biggest mass was localized beneath the occiput and right petrous bone (Fig. 1a, b). Multiple myeloma was suspected because of the skull base destruction and the mass observed.

Laboratory examination showed the following: hemoglobin 7.3 mmol/L (normal range: 8.0–10.5 mmol/L), leukocyte count $10.5 \times 10^9/L$ (normal range: 3–10), erythrocyte sedimentation rate (ESR) 42 mm/h, and C-reactive protein (CRP) 89 mg/L (normal: <8 mg/L). Following the results of the CT, additional blood tests were ordered: M-protein 4 g/L, IgG kappa, free kappa 35.3 mg/L (normal range: 3.3–19.4 mg/L). A ^{18}F -fluorodeoxyglucose (FDG)-positron emission tomography (PET)/CT of the body was performed for oncological analyses. It showed the metabolically active skull base lesion suspicious for malignancy but no signs of a primary tumor (Fig. 1c).

Initially, the patient was misdiagnosed as having probable myeloma. Magnetic resonance imaging (MRI) of the brain showed a big destructive process centered in the right midline of the apex of the petrous bone. It expanded to the clivus and the contralateral side of the corpus C1, with breakthrough in the right sphenoidal sinus. It connected with the mastoid and the middle ear. Further expansion was shown in the prevertebral soft tissues but also in the perivertebral space posteriorly as well as caudally around the vascular nerve strand via the jugular foramen and the hypoglossal foramen. It was suspicious for necrosis or abscess formation (Fig. 1d, e).

The differential diagnosis was wide and (apart from multiple myeloma and osteomyelitis) included nasopharyngeal carcinoma, metastasis, and inflammatory diseases. An ultrasound-guided puncture was performed in the paravertebral soft tissue, which showed *Streptococcus pneumoniae* as the causative agent. Treatment with intravenous penicillin 6 million units per

24 h for 6 weeks was initiated. Additional ear, nose, and throat history revealed that the patient also suffered from progressive hearing loss on the right side. He also reported to have had a history of ear infections and a draining right ear. Ear, nose, and throat examination showed a soft but intact external ear canal with a purulent otitis media on the right side. A paracentesis with tubes was performed. After 14 days of antibiotics intravenously, he was discharged and followed at the outpatient clinic. The headache improved but did not disappear; however, the otalgia vanished.

The mildly risen M-protein level was suspicious for monoclonal gammopathy of unknown significance. Two weeks after initiation of antibiotic treatment, blood tests showed the following results: leukocyte count $6.2 \times 10^9/L$, CRP 6 mg/L, and ESR 50 mm/h. Six months later, ESR was 20 mm/h. CT of the skull base was repeated after 3 months and showed improvement of the osteomyelitis of the skull base with a decrease in lytic abnormalities. In addition, improved air retention of the mastoid, middle ear, and sphenoidal sinus was observed (Fig. 1f).

Discussion

We report the case of a 70-year-old man with a history of diabetes who presented with chronic daily headache and right-sided otalgia that had been present for 6 months, who was found to have widespread skull base osteomyelitis which initially was misdiagnosed as myeloma.

There is some confusion on the terminology of temporal bone or skull base osteomyelitis as some authors also use “atypical osteomyelitis” and “central skull base osteomyelitis.” In the central skull base form, the sphenoid and/or occipital bones are affected. Others use “lateral temporal bone” and “medial temporal bone osteomyelitis” [1].

In our case, the osteomyelitis was located in atypical anatomical regions, namely the petrous bone, the clivus, and the contralateral side of the corpus C1, with breakthrough in the right sphenoidal sinus. There was no primary sphenoidal focus, mainly because of the clinical exam and the negative outcome of the FDG-PET/CT. Furthermore, in our case, there was no evidence of a malignant external otitis. *P. aeruginosa* is the most common causative pathogenic microorganism, although others have been identified, including fungal species [2]. *S. pneumoniae*, as in our case, as a causative agent is uncommon. Ears and teeth are the major sources of infection, but sinonasal infections, traumatism, surgery, and hematogenous spread of a distant infective focus have also been reported [3]. In our case, there were no signs of a dental source of infection.

Skull base osteomyelitis is a life-threatening condition. Severe complications are venous sinus thrombosis, meningitis, cerebritis, or abscess formation [4]. The main clinical features of skull base osteomyelitis are otalgia, facial pain, and headaches [5]. Also, (multiple) lower cranial nerve neuropathies can be seen. Treatment of skull base osteomyelitis is systemic antimicrobial treatment based on microbial culture of specimens [2]. Initial treatment should include broad-spectrum antibiotics which should cover *Pseudomonas*. Also, antifungal therapy should be considered [6]. Surgery has a limited role, being performed to get specimens or exclude malignancy.

Headache in combination with signs of skull base destruction can falsely point towards malignancy as a diagnosis. Especially as blood tests also suggest malignancy, as was the case in our patient, multiple myeloma was initially considered.

Patients presenting with persistent daily headache with a history of diabetes or otherwise immune compromised need appropriate clinical and radiological investigation, keeping the diagnosis of central skull base osteomyelitis in mind.

Statement of Ethics

The subject gave their written informed consent to publish their case.

Disclosure Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Jens van der Valk drafted the paper and final version and collected the data. Frank Treurniet collected the data and made the figures and legends. Jan Pieter Koopman contributed to the draft and approved the final version. Hille Koppen drafted the paper and the final version and collected the data.

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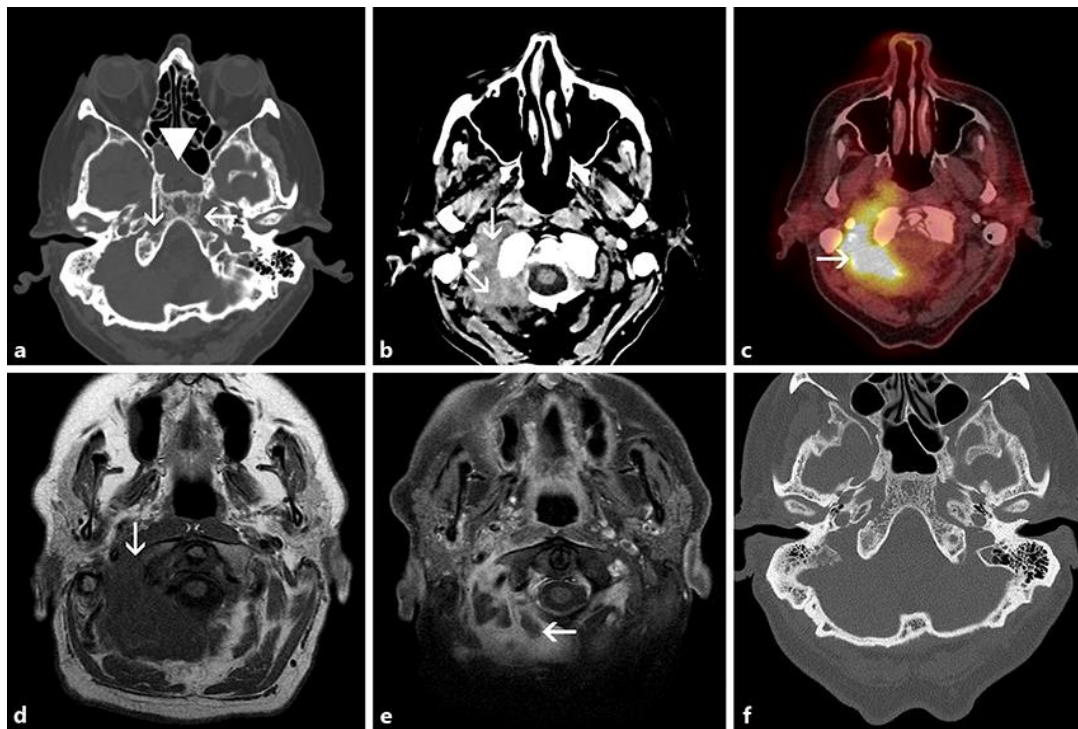


Fig. 1. **a** Axial bone CT shows destruction on both sides of the clivus (arrows) and mucosal thickening in the sphenoid sinus (arrowhead). **b** Axial nonenhanced CT shows a soft-tissue mass beneath the right side of the skull base (arrows). **c** Axial fusion FDG-PET/CT shows hypermetabolism of this tissue mass (arrow). **d** Axial MRI (T1-weighted, TSE) shows the extension of the tissue mass beneath the right occipital bone (arrow). **e** Axial MRI (T1-weighted, gadolinium-enhanced TSE) shows abscesses (arrow). **f** Axial bone CT 3 months later shows partial restoration of the skull base destruction, as well as air in right-sided mastoid cells.